AN INQUIRY

CONCERNING THE

NATURE AND CAUSES OF THE DISEASE DESCRIBED BY CRUVEILHIER

AS

ROGRESSIVE MUSCULAR ATROPHY.

A THESIS FOR AN ACT FOR THE DEGREE OF M.B. IN THE UNIVERSITY OF CAMBRIDGE.

BY

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THESIS.

That a portion of the cases of Progressive Muscular Atrophy has for one cause excessive involuntary use of the muscles atrophied, and that this involuntary use is due to the irritation on the peripheral nerves of poisoned blood circulating in them.

PRIOR to entering upon this thesis, for the sake of clearness, I will cite my clinical reports on two cases in point, which have recently fallen under my observation at St. Bartholomew's Hospital.

Charles Lodey, under Dr. Andrew, in Mark Ward of St. Bartholomew's Hospital from September 26th, 1873, until the end of December, 1873, gave the following history :—

Until about two years ago he was quite well.

About this time he was working in hot rooms, and had to go out of them into the cold and sometimes the wet, when in violent perspirations with but little clothing on. He drank rather

freely of beer. He complained of ill-defined pains in his limbs and back, which the doctor called rheumatic. During this attack, and towards the end of it he says he passed something like red sand in his water, and on one occasion being unable to pass his water, it was drawn off with a catheter. He then whilst at work was seized with violent cramps in his arms, forearms, and chest muscles, which continued for many hours. He says the muscles were as hard as marble, and the fingers drawn up tightly, and that the muscles of the right arm were harder than those of the left, and the chest muscles were cramped longer and more on the right side than on the left. On the cramps passing off he found he had lost much power in the affected muscles, and they then rapidly wasted, so that at the end of a very short time he found himself in the state in which he now is, and he says that he has neither improved nor got worse, sensibly, since. (I think fourteen days after the cramps.)

He has been twice under observation at St. Bartholomew's Hospital, and was in Guy's in July, 1873. At no time have any muscular twitchings or fibrillary tremors been noticed.

Guy's Hospital Registrations.

In Guy's Hospital he was thought to improve slightly under the Faradaic current. Loss of sensation to both currents in both arms. Muscles of right upper arm do not respond to continuous

current, those of the forearm do slightly to 60 Daniel cells. Interossei do not respond.

Present State.—Temperature 98·5°; respiration 18; pulse 68, fair volume, regular; face natural; skin warm; bowels regular.

Has considerable wasting of extensors and flexors of both forearms and muscles of hands, "mains en griffe." Right upper arm 8½ in. circumference; left is 9½ in. His right pectoral muscles are more wasted than the left, and he is right-handed. Some want of fulness of scapular muscles. Elsewhere his muscles are healthy, and his general health is good.

In this case—

I. The wasting followed directly upon the cramps (*i.e.*, excessive involuntary use) and in those muscles only which were cramped.

II. The cramps came at a time when there was evidence that his system was loaded with uric acid, which was probably the source of irritation on the nerves causing the cramps.

III. The rapidity of wasting was proportional to the excessive uses. Firstly, because the muscles remained violently contracted for some hours. Secondly, because those muscles wasted the most which were the most violently and the longest contracted—viz., those of the right side, although he was right-handed.

IV. His muscles quickly attained their maxi-

mum state of atrophy; he had no repetition of cramps, never any muscular twitching, and no progression of atrophy.

Francis Gowran, aged sixty, under Dr. Andrew, in Mark Ward, St. Bartholomew's Hospital, December 1st, 1873.

Was quite well until December, 1872, when he found a slight inability to write with a piece of chalk, after which (he fancied the metacarpo-phalangeal joint was growing out) the muscles of the ball of the thumb were observed gradually to waste. About the same time he had pains in his legs like rheumatism, and lasting about half an hour at night after a hard day's work, and this occurred only six or seven times. Although he has had twitching of muscles of the arms and legs to a considerable extent, he never noticed them in the arms until they were pointed out to him by a friend, but he had felt them in his legs. Fibrillary tremors have continued steadily during the past twelve months during waking and sleeping. Muscles have wasted steadily and quite gradually during the past twelve months.

From 1832 until the beginning of present illness he has been a policeman, dock labourer, and lastly, a night-watchman. Has always been much exposed to weather. Had two severe shakes—one a fall from a 10-foot wall in January, 1872.

Present History.—Muscles of both upper ex-

tremities wasted, especially the hands. Legs also wasted, and muscles of back; and in all these there are continually fibrillary tremors.

His temperature was constantly 98°, morning, afternoon, and night. Pulse 88. Weight 10 st. 3 lbs. Eighteen months ago he weighed 12 st.

From this I conclude that—

I. The atrophy was due to the excessive involuntary action.

II. The rapidity of atrophy was proportional to the excessive involuntary action.

III. It was developed in a man who has been much exposed to cold, one of the causes of poisoned blood.

IV. There was twitching in the wasting muscles.

Patients, especially if uneducated, are very unobservant. He did not notice the thumb wasting until he could not hold the chalk. He did not notice the fibrillary tremors and twitchings until they were pointed out to him; nor did he attribute the pains occasionally felt in the legs to cramp in the muscles, which they in all probability were. Therefore it is fair to assume that the wasting and twitchings were going on for some time before. And so I take it that in the cases where patients say they never had muscular twitching their opinion is worthless, as is also their opinion as to the date of the commencement of a gradual wasting, for they would date it from the time

they found themselves unable to perform some particular act.

The force of external circumstances acting upon the nerves of special sense of the scrivener, and being in due course modified by his brain, passes on to the motor nerves, and by them causes the frequent contraction of certain sets of muscles in his arms, alternating with periods of rest, and with but few exceptions each muscle contracting throughout its whole substance. When a muscle contracts, nearly all the blood in its veins is squeezed out, and during the subsequent relaxation is unable to return on account of the venous valves, and must be replaced by fresh blood from the arteries, the onward course of which is not now impeded by the presence of any blood in the veins. The more frequently these contractions alternating with relaxation occur, the more blood will pass through the muscle in question—that is, in this sense, the opportunities for nutrition of a muscle are proportional to the amount of use it undergoes. Now the muscles in the scrivener's hand profit by this opportunity up to a certain point, for thus far they steadily increase in size and power.

In every body and every part of a body is an inherent power of attaining, first, a maximum of development, and then of declining to its minimum of vitality, followed by death. Now if the functions of any part are called into play too

quickly, it will by so much the earlier exhaust that inherent power of self-nutrition and repair; thus we see that the hand of the scrivener, after remaining at its maximum state of development for some time, undergoes a premature atrophy and becomes useless—the muscles becoming little else than fat and fibrous tissue. The pathological explanation of this being that the muscular cells retain more and more of the fat which results from the splitting up of the albumen, which finally yields urea, kreatinine, kreatine, leucin, tyrosin, and sarco-lactic acid, and at the same pace loses the power of replacing itself by a new and strong muscular cell; and when the muscle has diminished in size, from loss of the true muscular substance, the fibrous tissue appears to be in great excess, for it formed the supporting network of a large organ when the muscle existed in full vigour, and has not diminished in proportion with the wasting of the muscle.

Carpenter's "Human Physiology:" on Urine.

Now if the scrivener had worked more moderately, he might have proportionally delayed the period of development of the atrophy, or avoided it till he attained the limit assigned by Nature.

In scrivener's palsy the wasting is proportional to excessive use, for

(*a*) It is commoner in men than in women, because men are more commonly scriveners than women.

Niemeyer on Scrivener's Palsy.

(*b*) Men in writing use more exertion, for they make thick down-strokes, and women's writing is of uniform thickness—in fact, little more than scratches upon the paper.

(*c*) When soft pens were used, scrivener's palsy was not noticed, for this disease came with the introduction of hard steel pens.

From which I conclude—

I. That the atrophy is due to excessive use, bringing on the natural termination prematurely.

II. That the rapidity of atrophy is proportional to the excessive use.

III. That the stimulation to contraction was caused by the necessities of his business.

IV. That it is a fatty degeneration of muscular tissue proper.

The case of the scrivener is the wasting following the voluntary excessive use of voluntary muscles induced by the unwholesome stimulation of living at high pressure, acting upon his mind. The two cited cases of muscular atrophy seem to have been caused by the unwholesome stimulation of the unhealthy blood acting either on the peripheral sensory nerves in the muscles or the nerve centres in the cord, and causing involuntary contractions of voluntary muscles. Now the difference between these cases is not so great as may appear at first sight, for let us inquire

1. Under what circumstances involuntary muscles contract normally, and

2. Under what circumstances voluntary muscles contract involuntarily.

The difference between voluntary and involuntary muscles is, that the former move by external circumstances, stimulating our special senses and the force passing through our intellects, and after being here modified according to what is called the temperament of the individual, passing on along the motor tract of nerves, and causing the contraction of various sets of muscles; or, in other words, bringing out a certain expression either of face, body, or words.

Hering's experiments.

The latter move by states of the body, as for example, when the blood becomes charged with carbonic acid, it stimulates the nerve centres of the respiratory muscles, and this stimulation is communicated along the nerves and causes the respiratory muscles to contract and draw a fresh volume of oxygen into the lungs for the aëration of the blood. Again the bile in the intestines stimulates the peripheral nerves there, and reflexly causes the peristaltic action of the intestines; also the presence of food in the stomach stimulates the peripheral nerves there, and causes the waves in it which occur during digestion.

We find also that the presence of fæces in the

rectum causes reflex contractions tending to expel them. Many other illustrations might be cited, all of which would demonstrate the involuntary motions caused by stimulating the peripheral nerves, which stimulation passing along the nerve to the centre, and not through the intellect, is then passed on through the motor nerve to the so-called involuntary muscles; and inasmuch as this stimulation or force is not modified by the intellect, the results of similar stimulation in different persons' involuntary systems are much more alike than the results of similar stimulations in different persons' voluntary systems. That is, the voluntary is only different from the involuntary system in that it possesses one more factor—*i.e.*, the modifying force of the intellect through which the prime force (or stimulation) passes, or a muscle acting reflexly is or may be the same as an involuntary muscle exactly. But the voluntary muscles, if stimulated locally, contract reflexly without the force necessarily passing through the intellect.

The blood varies in properties and composition at different times consistently with health—that is, in temperature, pressure, rapidity of current, and proportion of chemical constituents; and if these are altered so as not to be consistent with health, we have to all intents a poisoned state of the blood or poisoned blood. It is a very common thing for those who are subjects of known blood poisons to

have spasms of the muscles, cramps, or convulsions, as—

I. Convulsions occur in asphyxia where there is an excess of CO_2 in the blood and a want of oxygen.

II. In anæmia from bleeding also, convulsions are not uncommon, and here there is either an excess of water in the blood, or want of red corpuscles and consequently a want of oxygen. There is also violent peristaltic action of the intestines. Thus under the same set of circumstances we find both the voluntary and involuntary muscles acting similarly, that is to say, reflexly. Kussmaul and Tenner, New Sydenham Society.

III. Convulsions take place in persons who have an excess of urea in the blood, as in Bright's disease.

IV. Pyæmic patients are subject to epileptiform seizures.

V. Convulsions occur also in pregnancy.

VI. In eruptive fevers some of the convulsions may be explained by the reflex stimulation on the nervous system of the heated blood, and its otherwise altered or poisonous condition.

VII. In cases of snake-bites, where the blood contains an organic poison, the patients become delirious, and get convulsions.

VIII. Cramps, or muscular contractions, are very common in cholera; they may last as long as fourteen hours, and are of so violent a nature that Reynolds' "System:" Local Spasm.

muscles have been known to become ruptured. That the contractions in this instance are due to poisoned blood circulating in the limb is fairly well shown by the fact, that the local contractions cease after a short time, say in the leg, if the femoral artery be compressed.

IX. Strychnia in the blood causes violent muscular contractions.

X. Persons of gouty diathesis are very subject to cramps, manifesting themselves sometimes by evident muscular contractions of some severity, and at other times simply by pains in the muscles, which are subject to the contractions.

XI. Rheumatic patients also are frequently subject to cramps.

The eleven classes of causes enumerated above as giving rise to involuntary contractions of the voluntary muscles as a sequence to the presence in the blood of a known poison, are sufficiently numerous for the purposes of my argument, though others might easily be cited. I now pass to the more strictly physiological portion of my subject, and enumerate the four most common forms of abnormal change induced by the disease—that is to say, I pass from causes to effects.

Atrophy of a muscle might be due to—

1. The inherent power of nutrition being worn out by contraction or spasms of the muscle.

2. Thickening, or other alteration in the coats of

the capillary vessels which prevented the nutrition being carried on as it is in the normal state.

3. Altered condition of the nutrient fluids so that they are no longer capable of feeding a muscle.

4. Spasm of the vessels.

My reasons for believing the first proposition are gathered from the observations on the life of cells witnessed during the process of inflammation, for there we see that the more rapid the cell development, the more abortive are the cells produced, and the more likely too, to undergo degeneration, and the less capable of forming permanent tissues. Rindfleisch.

As regards the second proposition—thickening of the finer vessels might be brought about by abnormal states of the blood, such as hydræmia, gouty or rheumatic diathesis, diabetes, or the retention in the blood of too large quantities of its normal constituents, which is probably the real cause. So that the same blood poison which causes the involuntary action by irritation, causes that thickening of the vessels which is followed by consequent mal-nutrition.

Thickening of the sarcolemma would have the same effect, for in either case the nutrition is placed at a distance from the cells where its services for repair are required.

Thirdly, if the blood be continually at a low temperature, say even as slightly removed from the

normal as 97°, the same chemical products would either not be produced, or, if they were, would not be in such large quantities as if it were at 98·4°, although too delicate a difference to be detected by the rough methods now used. Now the blood in *Progressive* Muscular Atrophy cases, is usually rather low in temperature at the time when the atrophy is progressing.

In gout or rheumatism we find the patients subject to cramps and their blood-vessels are indisputably atheromatous—that is, there are changes in the walls of the blood-vessels, caused by inflammation brought about by the unwholesome stimulation of unhealthy blood.

Thus the three causes are much connected with one another, and may all be acting together, for if the inherent power of nutrition of the tissues is ever worn out, thickening in the vessels would accelerate it, and an altered state in the blood acts as a source of irritation to the blood-vessels, which causes thickening and change in them.

No. 4 is a possible, but very unlikely cause, and the effects which would follow are sufficiently obvious.

The first change in the muscular fibres in this disease is the disappearance of the transverse and then of the longitudinal striæ, and the next the appearance of granular segmentation, which becomes replaced by opaque granular substance within the sarcolemma. The fibres then contract, and become

S. Jacoub, "Leçons Chir. Méd."

filled with fat, which may remain or become absorbed.

Out of 110 cases there are records of 13 post-mortem examinations altogether, which are reliable for both the examination of muscular and nervous systems. Oppenheimer's and Meryon's cases were very carefully examined microscopically, and they found the spinal cord quite healthy. Cruveilhier, and Reade of Dublin, give 5 cases in which there was atrophy of the anterior roots of the spinal nerves, but, on the other hand, there are 5 cases equally carefully examined (of (1) Dr. Meryon's, "Med.-Chir. Trans.," vol. xxv. p. 23; (2) Landry, "Gaz. Méd.," No. 17, 1853; (3) Oppenheimer; (4) Virchow's "Archiv," Band viii.; (5) Laboulbène, "Union Méd.," Dec. 15, 1855) which show no alteration of the kind, although specially looked for. These indisputable facts prove that the disease is not due to a lesion of structure in the cord or anterior roots of spinal nerves. And that impressions are freely conveyed from the head to the muscles through the points in the cord and nerves, said by authors to be diseased, is evidence of absence of functional disease. Also the absence of all paralysis during life and the integrity of the intellectual functions, are evidence of absence of functional as well as of structural disease of the great nerve centres. I may therefore assert that atrophy of the nerves is not essen-

Roberts's Essay on Wasting Palsy.

(97, 98).

Trousseau on Muscular Atrophy.

tial, and therefore not primary. The only points in the disease which are constant, are atrophy of the muscles, a history of excessive voluntary use, as seen in 27 cases out of 110; or involuntary use of the muscles atrophied, as seen (in 35 out of 110 cases) in the form of muscular twitchings, fibrillary tremors, or cramps which may have existed in a great many others which were not examined at the time the muscles were wasting. In many cases the temperature of the blood is low at the time of the fibrillary tremors, and in the 110 cases recorded there is evidence of an abnormal state of the blood in 30 cases, and these are nearly all rheumatism, gout, or exposure to low temperature, stopping the excretions of the chemical compounds which had but just previously formed in the body by exercise.

When we find, in a post-mortem, muscles atrophied, and nerves connected with them also atrophied, we cannot, if really in search of the truth, say that the nerve was first diseased, and that this caused the muscles to undergo change. We must look to many observations on the constancy of the facts observed, and then we find the nerves are not always diseased. But if the muscles atrophy primarily, we might know *primâ facie* that the nerves would sooner or later become atrophied. And we find this to be the case. Although in some cases of disease of the supra-renal capsule, but

Niemeyer, vol. ii. p. 52.

not in all, there is an atrophy of the neighbouring plexus of nerves, we should not say that the disease of the supra-renal capsule was due to the atrophy of the nerves, but we are bound to argue the other way.

Taking these points into consideration, I conclude that the actual cause of the atrophy is the excessive action of the muscles which takes place during cramps, fibrillary tremors, or muscular twitchings; that this is in many cases caused by a poisoned state of the blood irritating the nerves; and that when the muscle is worn out, the nerves or nerve centres sooner or later atrophy, and undergo the changes so often observed.

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SLEEPLESSNESS